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Original Article

Linear dermatoses as a new, fascinating teaching topic with remarkable clinical findings

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ABSTRACT

Background: Skin diseases that take a linear distribution are a well-recognized group of skin problems such as linear morphea and inflammatory linear epidermal nevus. **Objective:** The objective was to gather all cutaneous diseases with linear dermatoses that primarily have a linear fashion, trying to classify them into well-recognized groups. **Patients and Methods:** This was a case-series, descriptive project where all patients with linear dermatoses were well evaluated during the period from 2014 to 2024. All demographic and clinical features were recognized and assessed. These diseases were confirmed by histopathological examination. **Results:** A total of 176 patients with linear dermatoses were observed and evaluated, with ages ranging from 4 to 45 years, with a mean of 31 years, with males (51.13%) and females (48.86%). The following skin diseases were observed: linear morphea in 39.20% of the cases. Encope de sabre (ECDS) morphea was detected in 43 (24.43 %) cases. Linear lichen planus was seen in 23.3%. All patients presented with non-itchy, linear, bizarre, pigmented, scarring atrophic lesions that were located on the face, neck, and axillae, while non-scarring, pigmented on the limbs. An inflammatory linear verrucous epidermal nevus was observed in 5.68% of the cases. All had linear lesions with scaly erythematous features. Epidermal nevus was detected in 2.27% of the cases, psoriasis in 1.70%, and lichen striatus in 1.13%. **Conclusion:** Linear dermatoses are a well-recognized group of skin diseases that have diagnostic importance and are commonly observed among patients with morphea, including linear morphea (39.20%) and encope de sabre (24.43 %), while linear lichen planus (23.3%) is the second most common cause. Other skin diseases are not common causes of linear fashion.

Key words: Linear dermatoses, Morphea, Lichen planus, ILVEN, Lichen striatus

INTRODUCTION

Various mechanisms of association that may induce skin diseases that are usually featured by diffusely distributed lesions show themselves in a certain linear pattern, or alternatively, they might distinctly spare such a linear pattern [1]. There are three such mechanisms involved with linear lesions such as Koebner phenomenon, reverse Koebner phenomenon, and Wolf's isotopic response [1,2]. Marks induced by scratching are usually linear in shape, koebnerized diseases primarily show themselves in a linear pattern [3]. The Renbök phenomenon designates the withdrawal of lesions when a different one appears, such as in a patient with psoriasis who experienced regression of psoriasis on the scalp concurrently with a patch of alopecia areata. Thus, a preexisting skin disease protects the patient from the manifestation of another skin problem [2]. The third mechanism, an isotopic response, is that a pathogenetically unrelated skin disease develops at the site of an entirely different, previously healed skin disease. To give an example, lichen planus may develop at the same site previously affected by herpes zoster [3].

Linear morphea is an inflammatory disease of the skin that may affect soft tissue with devastating functional and cosmetic impairment [4]. Recently, Sharquie et al. classified morphea into six variants, including ordinary, linear, encoup de sabre, generalized, mixed and lastly

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yellow morphea (xanthomatous morphea) [5,6]. It was a rare skin problem among Iraqis in the 1970s, yet the disease has increased in frequency since the 1980s and increased tremendously after 1991, and it is no longer a rare variety today [5]. Lichen planus pigmentosus (LPP) is considered a rare variant of lichen planus (LP) by most authors, considering their similar immunopathogenesis and occasional coexistence in some patients. LPP is characterized by a symmetrical, linear distribution of dark brown to gray or gray-blue, round or oval macules [7].

Inflammatory linear verrucous epidermal nevus (ILVEN), the classic "dermatitis" epidermal nevus pattern, is the most common and mainly presents as hyperkeratotic plaques with little scales, usually with a linear distribution following Blaschko's lines [8]. Epidermal nevi (EN) include a heterogeneous group of benign skin hamartomas. Usually, they are present at birth or in early childhood. Interestingly, EN often occurs in a whorled or linear pattern best described as *Blaschko* linear [9].

Linear porokeratosis is an uncommon variant of the disease. It is characterized by multiple papules and hyperkeratotic plaques in a linear pattern on the limbs. Lesions usually follow a Blaschkoid pattern [10]. Linear psoriasis is characterized by a linear distribution of psoriatic lesions along Blaschko's lines. True linear psoriasis in the absence of lesions elsewhere is extremely rare with only several cases reported in the literature [11]. Lichen striatus (LS) is a self-limiting linear dermatosis that mainly involves children aged 5 months to 15 years and is distributed with the lines of Blaschko [12]. On the other hand, Blaschkitis is a rare acquired inflammatory dermatosis, which follows the lines of Blaschko of unknown etiology and shares numerous similarities with lichen striatus [13].

Folliculitis decalvans is a neutrophilic primary cicatricial alopecia characterized by chronic inflammation, hair tufting, and follicular destruction leading to irreversible linear hair loss and permanent scarring of the scalp [14]. Granuloma annulare is a benign granulomatous disorder that may show anywhere on the body. It presents with a wide variety of clinical features. However, patients with a linear fashion, especially with finger involvement, are very rare [15,16].

This study aims to gather all skin diseases presented with linear dermatoses that have a primarily linear fashion, trying to classify them into well-recognized groups to be considered a new platform for teaching.

MATERIALS AND METHODS

This is a case-series, descriptive project where all patients with linear dermatoses were evaluated during the period from 2014 to 2024. A detailed inquiry about all demographic and clinical features was done and assessed carefully. Patient agreement for participation in the study was taken and informed consent was received by the participants or their parents. Patient privacy was considered, and their agreement to share and publish photos was received. Digital photography was done for the lessons. Histopathological examination was conducted as a confirmatory test for enquiry cases.

Ethics Statement

The Ethical Committee of College of Medicine, Tikrit University. The committee strictly adheres to "Standards and Operational Guidance for Ethics Review of Health-Related Research with Human Participants" and "Helsinki declaration revised 2013".

Members of the Scientific Research Ethical Committee: Prof. Dr. Wisam Al Dory; Asso. Prof. Ahmed Abdul Aziz; Asso. Prof. Dr. Abdul Sattar H. Abdullah; Asso. Prof. Dr. Osama Al Jubory.

RESULTS

A total of 176 patients (Table 1) with linear dermatoses were observed and analyzed, with their ages ranging from 4 to 45 years, with 90 (51.13%) males and 86 (48.86%) females. The following skin diseases were recorded: linear morphea (Fig. 1a) in 69 (39.20%) cases, ages ranging from 4 to 45 years, with a mean of 28 years, with 41 (59.42%) females and 28 (40.57%) males. The limbs were the most commonly involved sites, especially the lower limbs. Although the lesions began as pigmented patches, the inflammatory process was more rapid and progressive and quickly changed into atrophy and sclerosis. It was usually unilateral, but one case with linear morphea was seen in both upper and lower limbs. In late cases, there was even atrophy of the underlying fascia and muscles, and even bones in some patients.

Encope de sabre (ECDS) morphea (Fig. 2a) was detected in 43 (24.43 %) cases, with their ages ranging from 4 to 32 years, with a mean of 20 years, 34 (79%) cases under 18 years of age, with 30 (69.76%) females and 13 (30.23%) males, with a female-to-male ratio of 2.3:1. The lesions also began as pigmented, non-scaly

Figure 1: Showing linear morphea of the left lower limb (a). Linear inflammatory epidermal nevus (b). Linear psoriasis (c). Linear granuloma annulare of the forearm (d). Linear Blaschkitis affecting the left arm, shoulder, chest, and neck (e).

Figure 2: Showing different linear facial skin lesions. Encope de sabre of the face (a). Linear lichen planus of the face (b). Epidermal nevus (c). Linear porokeratosis of the face (d). Chronic folliculitis decalvans of the scalp (e).

Table 1: The clinical, demographic characteristics and frequencies of linear dermatoses

Site	Clinical Lesion	Sex	Age (Yrs.) (Mean)	Dermatosis (%)
Lower limbs	Pigmented patches atrophy & sclerosis	59.42% females	4–45 (28)	Linear morphea (39.20%)
Frontoparietal scalp, para-median forehead	Indurated depressed sclerosed	69.76% females	4–20 (32)	Encope de sabre (24.43%)
Face, neck, & axillae	Bizarre pigmented atrophic	82.9% males	20–60 (37)	Linear lichen planus (23.3%)
Upper limbs, trunk	Scaly erythematous	80% males	4–12 (8)	ILVEN (5.68%)
		75% males	6–25	Epidermal nevus (2.27%)
		100% females	20–40	Psoriasis (1.70%)
Upper limbs		Male	10–12	Lichen striatus (1.13%)
		Male	25	Folliculitis decalvans (0.56%)
		Female	25	Porokeratosis (0.56%)
		Female	30	Granuloma annularae (0.56%)
Arm, shoulder, chest, neck	Dermatomal papules	Male	29	Blaschkitis (0.56%)

patches that, over time, become indurated sclerosed areas. It most commonly involves the frontoparietal scalp and/or the para-median forehead, and to a lesser extent the face. These lesions might eventually end with depressed, sclerosed, pigmented bands and even cause atrophy of the underlying bone. When the lesion involved the scalp, there was often atrophy of the underlying bone and scarring alopecia. These scary areas began as moth-eaten sites and coalesced together to end with big, scarred patches. All cases of ECDS presented unilateral involvement of the scalp or face, and none of them had bilateral skin involvement.

Inflammatory linear verrucous epidermal nevus (Fig. 1b) was observed in 10 (5.68%) cases, 8 (80%) males and 2 (20%) females; their ages ranged from 4 to 12 years, with a mean of 8 years. All had linear lesions with scaly, erythematous features involving the lower and upper limbs and some extending into the trunk and face.

Linear lichen planus (Fig. 2b) was seen in 41 (23.3%) cases, with their ages ranging from 20 to 60 years, with a mean of 37 years, 34 (82.9%) males and 7 (17%) females. All patients presented with non-itchy, linear,

bizarre, pigmented, atrophic lesions on the face, neck, and axillae in 37 (90.2%) cases and with non-scarring pigmented on the limbs in 4 (9.76%) cases.

Linear psoriasis (Fig. 1c) was seen in 3 (1.70%) cases; their ages ranged from 20 to 40 years and were all females.

Epidermal nevus (Fig. 2c) was detected in 4 (2.27%) cases; their ages ranged from 6 to 25 years, they were 3 (75%) males and 1 (25%) female.

Lichen striatus was observed in 2 (1.13%) male children; their ages were 12 and 10 years and both cases were on the upper arms.

Granuloma annularae (Fig. 1d) was seen in a 30-year-old female (0.56%).

Porokeratosis (Fig. 2d) was observed in a 25-year-old female (0.56%).

Blaschkitis (Fig. 1e) was observed in a 29-year-old male patient (0.56%) with a typical linear distribution involving the left arm, shoulder, chest, and neck. Histopathology showed a dense inflammatory reaction in the epidermis and dermis, with an intraepidermal blister (Fig. 3a and 3b).

Folliculitis decalvans (Fig. 2e) was observed in a 25-year-old male (0.56%).

DISCUSSION

Linear lesions in dermatology are fairly common cutaneous complaints in daily clinical work. However, the clinical picture can be deceptive, which puts dermatologists in a diagnostic dilemma. Intuitive analysis of the type and distribution pattern of skin lesions by distinguished dermatologists is the first step in making a list of possible differential diagnoses. The subsequent diagnostic workup, including imaging studies, histopathological assessment, and laboratory tests, either supports or excludes the diagnosis [17].

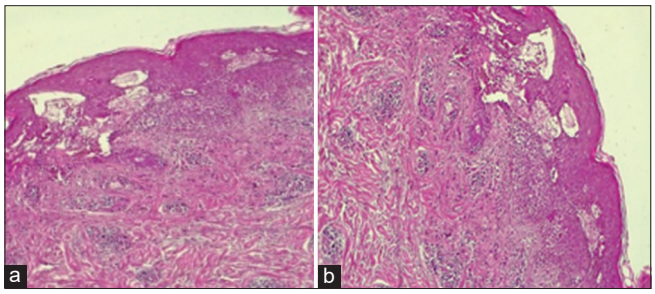

Figure 3: (a and b) Histopathology of Blaschkitis showing a dense inflammatory reaction of the epidermis and dermis, with an intraepidermal blister (H&E; 10x, 40x).

Numerous skin diseases present a neither even nor random distribution but are characterized by typical predilection sites. Some skin diseases occur in an even more localized pattern, as they follow certain linear forms. These patterns can be arranged into four major groups, Langer lines, dermatomes, Blaschko lines, and the exogenous patterns, as well as additional patterns associated with one of the major groups as the Christmas-tree pattern and the stretch-mark pattern.

Moreover, differential diagnostic considerations must also involve cutaneous disorders that usually show themselves with diffusely distributed lesions or as a rash, as they may adopt atypical patterns as a result of different mechanisms [17].

This new case series uniquely showed 176 patients with linear dermatoses; their ages ranged from 4 to 45 years, with an almost even male-to-female ratio of 1.04:1. The following skin diseases were seen in decreasing order (Table 1).

Linear morphea in 39.20% of the cases, more frequent in females (59.42%). The limbs were the most commonly involved sites, especially the lower limbs. This concurs with a study by Christen-Zaech, who found that, among 136 cases, most cases of morphea were linear, especially on the extremities, and were also more frequent in females than males (female-to-male ratio of 2.42:1) [18].

The morphology and predominance of morphea lesions was similarly mentioned in a previous study by Sharquie et al., who interestingly, reported that the lesions started as pigmented patches, but the inflammatory process was more rapid and progressive and quickly changed into atrophy and sclerosis. These histopathological findings were also correlated with the aforementioned study. It was usually unilateral, but one case with linear morphea was seen in both upper and lower limbs. In late cases, there was even atrophy of the underlying fascia and muscles, and even the bones in some patients [5,6].

Encope de sabre (ECDS) morphea was detected at relatively high rates (24.43%) and at young ages (4 to 32 years), with 79% of the cases under 18 years of age, with a female-to-male ratio of 2.3:1. The lesions

also began as pigmented, non-scaly patches that, over time, became indurated, sclerosed areas. It most commonly involved the frontoparietal scalp and/or the para-median forehead, and to a lesser extent, the face. These lesions might eventually end with depressed, sclerosed, pigmented bands and even cause atrophy of the underlying bone. When the lesion involved the scalp, there was often atrophy of the underlying bone and scarring alopecia. These scary areas had begun as moth-eaten sites and coalesced together to end with big, scarred patches. All cases of ECDS presented unilateral involvement of the scalp or face, and none of them had bilateral skin involvement. These findings were closely similar to a previous study by Sharquie et al. [5].

Linear lichen planus was relatively common in this series (23.3%) and was a disease of males (82.9%), which was in agreement with a study by Sharquie et al., where there was a male predominance, and all cases displayed linear sclerosing pigmented atrophic bizarre lesions of the face, while others were found to have linear lichen planus on the limbs, scalp, and penis [5]. Also, Dhar et al. reported linear lichen planus associated with mild itching in 27–62% [8], while all our cases presented with non-itchy linear bizarre pigmented atrophic lesions on the face, neck, and axillae in 90.2% of the cases and non-scarring pigmented on the limbs in 9.76%. There were similar sites of predilection between these two studies, with comparable results in terms of sex and age of patients, although they reported much younger ages of five years.

The present report agreed with a study by Tseng, where ILVEN occurred most commonly in children on the lower extremities and buttocks, while the involvement of the trunk, upper extremities, and genital area was possible. Surprisingly, the majority of our patients were male, 80%, which was in contrast to Altman and Mehregan diagnostic criteria of ILVEN where the female-to-male ratio was 4:1 [19].

Epidermal nevus was rarely reported in this trial as seen in 2.27% of the cases; their ages ranged from 6 to 25 years, 3 (75%) males and 1 (25%) females. These findings are lower than mentioned by another study that described epidermal nevi in 10% of linear dermatoses. This difference was probably due to different settings of the studies and different sample sizes (176 versus 100 patients). The current study obviously showed that the lesion is more frequent in males (75%), whereas there was no statistically significant correlation with sex in the Iranian study [20].

Lichen striatus in the present work was observed in 2 (1.13%) males, 12 and 10 years old, and both on the upper arms. While another study found 29 cases with lichen striatus with a mean age of 7.8 years when first presented. The lesions followed Blaschko lines in all, except three lesions were arranged along the axial lines of Sherrington [21]. In contrast to another retrospective study by Mendiratta, who described records of 30 patients with lichen striatus; 70% were females, while our cases were males. The most common age group affected was less than 4 years, which is younger than our patients [22]. In contrast to a recent study, it was the most common presentation among 90 patients with linear dermatosis [23].

Folliculitis decalvans were seen in one patient with classical features of the disease. Although it is a common cause of chronic folliculitis, it seems to be rarely presented in a linear fashion, as seen in the present work [24].

Although few case reports on typical Blaschkitis have been discussed in the literature, and no large studies are to be found, we reported a single case of typical Blaschkitis in a 29-year-old male patient with classical dermatomal distribution involving the left arm, shoulder, chest, and neck. The rarity of this disease concurs to Grosshans, who described the disease as a rare form of BLAISE in adults [13]. Also, it is comparable to another case by Fernando who reported a 49 year-old-man with multiple erythematous and hyperpigmented papules arranged in a linear Blaschkoid manner involving the lower abdomen. Although the age of our patient was younger and the site differed, both cases were males [25].

CONCLUSION

Linear dermatoses are a well-recognized group of skin diseases that have diagnostic importance and are commonly observed among patients with morphea, including linear morphea in 39.20% and encope de sabre in 24.43% of the patients. Linear lichen planus was the second most common cause. Meanwhile, other skin diseases are not commonly present in a linear fashion such as inflammatory linear verrucous epidermal nevus, epidermal nevus, psoriasis, lichen striatus, folliculitis decalvans, porokeratosis, granuloma annularae, and blaschkitis. This study may be considered a new platform for clinical teaching.

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Statement of Human and Animal Rights

All the procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the 2008 revision of the Declaration of Helsinki of 1975.

Statement of Informed Consent

Informed consent for participation in this study was obtained from all patients.

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